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Biliary atresia with aneurysmal dilatation of hepatic artery: A rare anomaly

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ABSTRACT

INTRODUCTION: The coexistent biliary atresia with aneurysmal dilatation of hepatic artery is a rare association. To know these anomalies will avoid many per operative complications. It is also important to mention that these children require liver transplantation in the long run then these vascular anomalies become more relevant.
PRESENTATION OF CASE: A four month old male child presented with features of biliary atresia. On exploration a cystic expansile mass was detected beneath thread like common bile duct. Subsequent aspiration and studies proved it to be aneurysmal dilatation of hepatic artery.
DISCUSSION: With biliary atresia many vascular and cardiac malformations have been described but aneurysmal dilatation of hepatic artery is a rare association. These anomalies may have impact on aetiopathogenesis of biliary atresia and also future liver transplantation.
CONCLUSION: Awareness of rare association of hepatic artery aneurysm with biliary atresia will help in understanding aetiopathogenesis of biliary atresia and planning liver transplantation in such cases.

1. Introduction

The association of biliary atresia with many congenital anomalies is a well established entity.[1] Portal vein anomalies and cardiovascular malformations are frequently associated with biliary atresia cases.[2] 20% of biliary atresia have been associated with other congenital anomalies.[3] The Japanese Biliary Atresia Registry (1989) have reported associated congenital anomalies in 114 cases out of 626 cases of biliary atresia. In a study at King's college London vascular malformations form 19.5% of all associated malformation in biliary atresia cases. The common vascular anomalies reported are anomalies of portal venous system, predoudenal portal vein, absent inferior vena cava and cardiac malformations. These associated anomalies in these patients make the prognosis poor.

The association of aneurysmal dilatation of hepatic artery with biliary atresia has not been reported yet in English literature.

2. Case report

A four-month-old male child presented with history of jaundice since birth. History of passage of clay colored stool and high colored urine was present. On physical examination, baby had severe jaundice with mild hepatosplenomegaly. Blood picture revealed leucocytosis, with conjugated hyperbilirubinemia (15.6 mg%). There was elevated level of serum transaminases, alkaline phosphatase and gamma glutamyl transpeptidase. Ultrasonography did not detect a gall bladder and extra hepatic biliary apparatus. There was no dilatation of intrahepatic biliary radicles. However a cystic swelling of size 5.5 cm × 6.5 cm was found in subhepatic area which on color Doppler supplementation showed whirling color flow pattern in it suggesting it to be aneurysm. It had its origin from the hepatic artery.

HIDDA scan after three days of phenobarbitone therapy suggested no secretions of tracer material into the gut. On laprotomy, atretic gallbladder with fibrous thread like extra hepatic biliary apparatus was found. A swelling was noted in the subhepatic space. This swelling was expansile in nature and was continued with hepatic artery proximally (Fig. 1). A 10 cc syringe was put into the swelling and bright red blood was aspirated (Fig. 2).

Liver biopsy was done. Histopathological report revealed bile duct proliferation, portal tract inflammation and extensive fibrosis suggestive of cirrhosis. Modified Kasai portoenterostomy was done in this case. In the follow up period, patient received ursodeoxycholic acid and there was improvement in serum bilirubin level though it did not came to basal level.

3. Discussion

Biliary atresia has been associated with a spectrum of associated malformations. These include malformations like jejunal,

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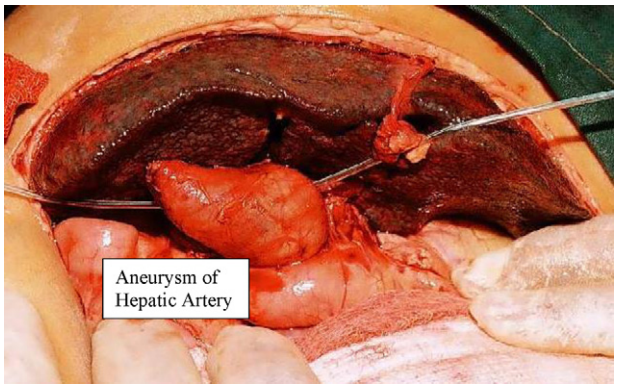

Fig. 1. Aneurysmal dilatation of hepatic artery in biliary atresia.

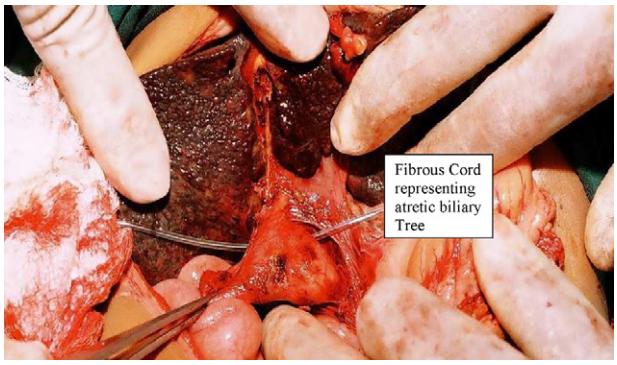

Fig. 2. Aneurysm of hepatic artery with fibrotic cord like structure of extra hepatic biliary apparatus.

duodenal and esophageal atresia, malrotation, annular pancreas, situs inversus, polysplenia, asplenia, cleft palate, polycystic kidney and talipes equinovarus. Vascular anomalies include portal vein malformations like predoudenal portal vein, cavernomatus malformation and agenesis of portal vein. Cardiac anomalies include ventricular septal defect, dextrocardia, immotile cilia (Kartagener's syndrome) and absent inferior vena cava. Many theories have been propagated for explaining the origin of biliary atresia. These include genetic causes,[4] bile duct canalization, and vascular accidents.[5] Cunnigham and Sybert has reported that in the causation of biliary atresia, a genetic susceptibility with environmental influence may be responsible.[6] Anatomical structural abnormalities has also been described like long common pancreatico-biliary channels in 28 cases reported by Chiba et al.[7] Vascular insult with environmental influence may be associated reason for biliary atresia including aneurysmal dilatation of hepatic artery.

Aneurysmal dilatation of hepatic artery as a cause of cystic dilatation during dissection in extra hepatic and portal area may be another differential diagnosis other than coexistent choledocal cyst.[8–10]

Ultimately in the long run these cases require liver transplantation. For this reason also, knowing this rare vascular anomaly may be of some help.

4. Conclusion

To know biliary aresia association with aneurysmal dilatation of hepatic artery is helpful in diagnosing and planning treatment of these patients.

Conflict of interest statement

None.

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Ethical approval

Obtained.

Author contributions

Bindey Kumar contributed for study design, Zaheer Hasan contributed for data collection, Prem Kumar and Utpal Anand contributed for data analysis, Rajiv Priyadarshi and Neelam Sinha contributed for writing.

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